

**Item S1: Distribution of mutation strength across pattern groups for *PKD1* participants**

**Table a: Study A (N=349 *PKD1* participants with available mutation strength)**

		Progressor & Linear (n=228)	Progressor & Nonlinear (n=77)	Nonprogressor (n=44)	p
Mutation strength	Truncating (MSG1)	146 (64.0%)	51 (68.0%)	14 (43.8%)	0.05
	Non Truncating (MSG2)	48 (21.1%)	17 (22.7%)	8 (25.0%)	
	Non Truncating (MSG3)	34 (14.9%)	7 (9.3%)	10 (31.3%)	

**Table b: Study B (N=331 *PKD1* participants with available mutation strength)**

		Progressor & Linear (n=273)	Progressor & Nonlinear (n=42)	Nonprogressor (n=16)	p
Mutation strength	Truncating (MSG1)	196 (71.8%)	25 (59.5%)	5 (31.3%)	< 0.01
	Non Truncating (MSG2)	50 (18.3%)	10 (23.8%)	5 (31.3%)	
	Non Truncating (MSG3)	27 (9.9%)	7 (16.7%)	6 (37.5%)	

MSG: Mutation strength group as defined by Heyer CM, Sundsbak JL, Abebe KZ, et al.. *J Am Soc Nephrol.* 2016; 27(9):2872-2884: MSG1 are truncating mutations, MSG2 are strong nontruncating mutations, whereas MSG3 are weak nontruncating mutations.